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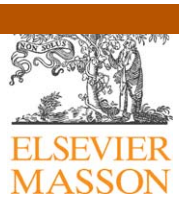

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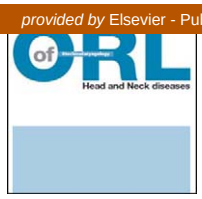

IMAGES, QUESTIONS AND ANSWERS

Congenital tumefaction of the medial canthus

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Clinical history

A six-week-old girl was referred for subcutaneous tumefaction facing the left medial canthus. The tumefaction had been present since birth, without evolution. Other than intermittent snoring, the child was asymptomatic.

Clinical examination found 15 mm bluish subcutaneous tumefaction, fluctuating under palpation, well-contoured and pain-free (Fig. 1). Eyeball, palpebral conjunctiva and lacrimal meatus examination was normal. Massage round the lacrimal sac induced no meatal effusion. Left nasal cavity examination, on the other hand, found a bluish cyst under the inferior concha (Fig. 2). Otherwise, clinical examination was normal.

To refine diagnosis, you asked for a maxillofacial CT-scan (Figs. 3 and 4).

Figure 1 Photograph of patient.

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Figure 2 Endonasal view (left nasal cavity).

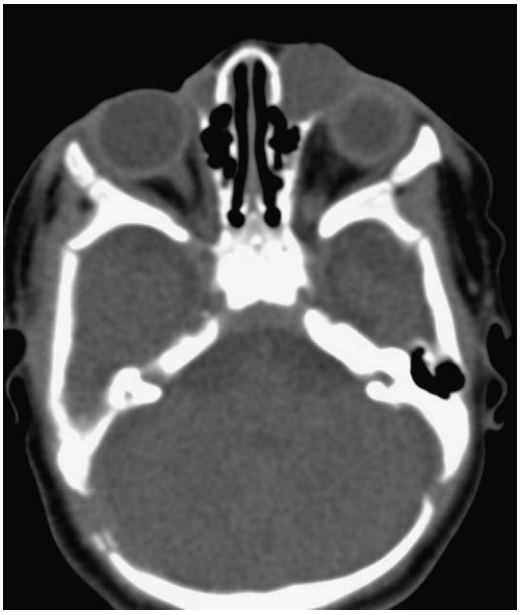

Figure 3 Maxillofacial CT: axial slice without contrast injection.

Figure 4 Maxillofacial CT: coronal slice without contrast injection.

Questions

Question 1

What are your diagnostic hypotheses based on the clinical examination?

Question 2

Describe the CT images (Fig. 3, Fig. 4).

Question 3

What is your diagnosis?

Question 4

What are the treatment options in this pathology?

Question 5

Which option do you choose, and why?

What are your diagnostic?

Answers

Answer nº 1

What are your diagnostic hypotheses based on the clinical examination?

- dacryocystocele;
- meningocele, meningoencephalocele;
- hemangioma, lymphangioma;
- sinus mucocele;
- malignant tumor (glioma, rhabdomyosarcoma).

Answer nº 2

Describe the CT images
The maxillofacial CT shows: an isodense, rounded, homogeneous and well-contoured lesion at the left medial canthus. It measures 15.6 × 15 mm in the axial plane and is 15.9 mm high. There is also left lacrimonasal duct dilation, prolapsed in the left nasal cavity under the inferior concha.

On the right side, there is slight dilation of the lacrimonasal duct and lacrimal sac.

Answer nº 3

What is your diagnosis?
Congenital dacryocystocele: cystic dilation of the lacrimal sac and lacrimonasal duct.

Answer nº 4

What are the treatment options in this pathology?
The treatment options are the following:

- lacrimal sac massage and clinical surveillance;
- lacrimal pathway probe/intubation;
- external surgical drainage;
- endonasal surgical drainage (marsupialization).

Answer nº 5

Which option do you choose, and why?
We suggest endoscopic endonasal drainage: i.e., marsupialization of the dacryocystocele (cyst opening and large excision of walls).

Surgery is justified as the risk of infectious and respiratory complication in dacryocystocele is high. Failure rates in conservative treatment (massage, probing) are elevated. Endoscopic drainage is more straightforward and less risky than external drainage. It is at present the attitude of choice for congenital dacryocystocele.

Comments

Dacryocystocele is a cystic dilation of the lacrimal sac and lacrimonasal duct secondary to simultaneous distal and proximal impermeability of the excretion system. It is induced by persistence of embryonic membranes in Hasner's and Rosenmüller's valves. First described in 1933 by Duke-Elder, it is a rare pathology with a clinical incidence estimated at 0.08%. According to a recent study, however, the radiological incidence of congenital dacryocystocele is higher, at about 0.7% [1].

Clinically, diagnosis is founded on an aspect of bluish cystic lesion in the medial canthus region, but 65% to 76% of dacryocystoceles present as acute dacryocystitis [2,3,4]. Series comprising endonasal examination and/or systematic imaging found associated cystic prolapse of the lacrimal duct in 60% to 96% of cases [1,4,6]. This endonasal component explains why congenital dacryocystocele can manifest as neonatal respiratory distress, even when unilateral.

The incidence of bilateral forms is probably underestimated. Clinically, bilateral forms are found in 8% to 27% of cases, whereas their radiological incidence is estimated at 42% [1,2,4,5].

Imaging can rule out differential diagnoses of an aspect of medial canthus tumefaction. It can also explore for any infraclinical contralateral lesion. Currently, maxillofacial CT, with or without contrast enhancement, is the reference examination.

Treatment for congenital dacryocystocele remains controversial, as spontaneous drainage may occur: the mean rate of spontaneous resolution is 23% [2,5]. The high risk of infectious complications, however, means this attitude is justifiable only over the short-term (one to three weeks). Lacrimal pathway probing or intubation showed variable success according to the series. In practice, it appears effective when there is no superinfection or endonasal cyst [2,3,5]. Endonasal marsupialization is a simple surgical procedure, and can be performed using cold instruments or laser. To date, no complications have been reported and recurrence after marsupialization is exceptional. It is at present the treatment of choice for congenital dacryocystocele.

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